SOME POST-HEMIPLEGIC DISTURBANCES OF MOTION IN CHILDREN.

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SOME POST-HEMIPLEGIC DISTURBANCES OF MOTION IN CHILDREN.[1]

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In a paper[2] which I had the honor of presenting to this Association a year ago, I spoke of the frequent occurrence of post-hemiplegic disturbances of motion in cerebral infantile paralysis. Certain forms of these disturbances have been carefully studied and are well known, among them being athetosis and post-hemiplegic chorea; other forms, such as ataxia, post-hemiplegic paralysis agitans, and post-hemiplegic pseudo-sclerosis, are less common. It is not my purpose, however, to speak in detail in this paper of these various forms of post-hemiplegic disturbance of motion. The exhaustive treatise of Greidenberg[3] renders such a task unnecessary, but it may be well to cite for reference the elaborate classification which he makes of these phenomena — the most thorough schedule that has yet appeared.

[1] Read before the American Neurological Association at the First Triennial Congress of American Physicians and Surgeons, 20th September, 1888.

[2] Hemiplegia in Childhood. Journal of Nervous and Mental Disease, August, 1887.

[3] B. Greidenberg. Ueber die posthemiplegischen Bewegungsstörungen, Archiv f. Psychiatrie, xvii. 131, 1886.

- Contractures
  - Apoplectic
    - Spasms
      - Clonic.
      - Tonic.
      - Intermitting.
    - Muscular rigidity.
  - Early. Paralytic, passive, temporary.
  - Late
    - Constant, continuous, fixed,
    - changeable (latent).
- Increased tendon reflexes.
- Associated movements.
- Tremors
  - Reflex—clonus.
  - Essential
    - Tremor proper.
    - In the form of paralysis agitans.
    - In the form of disseminated sclerosis.
- Hemichorea
  - Constant.
  - On intended movement — disturbance of co-ordination (hemiataxia).
- Athetosis.

Mixed forms in different combinations.

The following cases present a form of post-hemiplegic disturbance of motion which, on analysis, can be made out to be a combination of some of the forms in Greidenberg's table, but it is a form which has not yet been thoroughly described.

Observation I. George E., three and a half years old. First seen June 15, 1887. History of epilepsy in mother's family, and his mother herself is epileptic, having had two convulsions during pregnancy. He is the first child; was born after an easy labor of twelve hours' duration, without the use of instruments, the head presenting. No history of injury. He was a backward child, and did not sit up until the age of two. At present he can stand and walk a step or two with support, but usually does not attempt it, getting about by creeping. He has had one attack in the night, probably convulsive, of which no details could be obtained. Ever since he began to use his limbs his mother has noticed that he could not use his left arm well, and that his right leg was not as good as the left. He is said to be bright, to be able to sing with fair correctness, to repeat little verses, and to have a good memory. The speech, however, to a stranger

is quite indistinct. He breathes through the mouth, drools constantly, and has some slight difficulty in swallowing. He is subject to colds, as the clothing over his chest is constantly wet from his drooling. The appetite is dainty, and he has a delicate stomach. He still soils himself occasionally. The child is well developed, with no deformities, the two sides being symmetrical. The head is nineteen and three quarter inches in circumference. The left leg and arm are held rather rigidly, and there is inco-ordination when he tries to use the left hand, the movements being awkward and jerky, but there are no involuntary movements when the arm is at rest, with one exception. When he makes a movement elsewhere, there is often an associated movement of the left arm. The arm is rotated inwards, extended at the elbow, flexed and pronated at the wrist, and flexed at all the phalangeal joints; Individual movements of the arms, hands, and legs seem fairly well done, except (dorsal) extension of the right foot. Passive (dorsal) extension of the left foot reveals a slight constant rigidity. On attempting to walk, he walks with both feet inverted, and the right leg at times gives out. The left forefinger is usually kept clenched in the palm. Sensation, reflexes, and electrical reactions showed nothing unusual. There was nothing abnormal discovered in the movements of the face and eyes. He was referred to Dr. F. H. Hooper, who found a narrow, high-arched palate, and a mass of adenoid vegetations in the naso-pharynx, which were removed, relieving the drooling considerably. In September he had a convulsion, which his mother attributed to indigestion. In October his general health was better, and he got about better. In October there was considerable inco-ordination of

the left hand and a little of the right. The left hand was fairly strong. The gait was unsteady, the right foot being twitched after him. The associated movement above described persisted, and he still drooled somewhat. He crept rapidly, jerking his legs under him, and throwing his feet up in the air. January 4th, 1888, the condition was practically unchanged. He was beginning to walk a little, and his movements were less irregular and stronger. The inco-ordination and associated movements persisted. Some months later I received a letter from the family physician, telling me that the child had had more convulsions, still attributed to indigestion.

Observation II. Grace S., seven years. First seen May 11, 1887. Family history negative; first child. The labor was protracted and difficult, lasting from Tuesday until Friday. Forceps were used; the head after delivery was out of shape, and for some time there was a great question about resuscitating her. Her physician told me that she was nearer dead than any child he ever saw that recovered. She was always a large, fat child. At six months she had diarrhœa, and after that she could not sit up like other children. She sat alone at two, crept at three and a half, talked at four, and began to walk alone at five. About two years ago she began to have attacks at night, in which she became rigid, moaned, and frothed at the mouth, the arms twitched, and the right arm shook. After this she could not keep still. She never had more than twelve of these attacks. Her left side has always been the most affected, and she can use the left arm but little. When she does not feel well, the left hand will sometimes open and shut involuntarily. She is reported to be unusually bright in many

ways, to have an excellent memory and a large "bump of order." She notices quickly, and likes to help about the house. Lately, for no reason that she can give, she cries when left alone. She now sleeps well, but until she was three or four years old she slept very little, waking and screaming, apparently from pain. She talks a great deal, although her speech is nasal and very indistinct, but she understands what is said, and never uses the wrong word. She drools constantly, and takes cold easily. She breathes through the mouth, which is constantly kept open. She can swallow only liquids and soft food. When about five years old she had massage, after which she began to walk. The child is well-nourished and well-developed, the left arm being a little larger than the right. The left parietal region is more prominent. There is right external strabismus. The open mouth and the constant drooling give the child the appearance of an idiot. The right disk is smaller, and there is a lack of pigment, but no vascular changes. She can move the left arm but little, and the hand hardly at all, and the movements of the right hand are uncertain and not co-ordinated. The left arm is rigid, and kept rotated inwards and carried a little backwards, hyper-extended at the elbow, flexed and pronated at the wrist, and the fingers flexed over the thumb, which is between the fore and middle fingers. This rigidity relaxes somewhat at rest, but is heightened by any attempt at motion of either arm, or by excitement. Motion of the left hip and knee is fairly good; at the ankle there is slight motion, but the foot is held with some rigidity in equino-varus. The gait is peculiar; she walks on the left toe, the foot being inverted, and the left leg is weaker, giving out occasionally; the right

foot goes first, and the left leg is dragged after it, crossing the right, while the body is thrown forward at the hips, and progress is toward the right. On exertion or excitement there is also some rigidity of the right leg. She uses the right hand very slowly and awkwardly to pick up things, and the effort brings on associated movements in the left side,— either the spasm of the arm above described, or a similar spasm, except that the hyper-extension of the elbow is replaced by semi-flexion, or the hand is opened. The toes of the left foot are also extended (dorsally) and the inner edge of the foot is turned upwards — a form of associated movement to which Strümpell[4] has called attention. There is also great inco-ordination of the left hand, which she cannot open herself; but at times it opens on associated movements. When opened passively, the hand shuts slowly. There is less loss of power than inco-ordination. The tongue is protruded with difficulty. The knee jerks are little, if at all, exaggerated; there is no ankle clonus; the other reflexes and the electrical reactions show nothing unusual. As in the former case, Dr. Hooper found adenoid growths, which he removed, causing a marked improvement in the drooling, speech, swallowing, and general expression; the child ceased to look like an idiot. Massage and light gymnastics have aided the other conditions somewhat.

Observation III. M. L., female, six. Seen with Dr. Boland, of South Boston, October 7, 1886, and January 22, 1887. No nervous heredity could be discovered, and the family history, so far as known, is good. The child has been in the charge of a very intelligent woman, who has observed her with great

[4] A. Strümpell. Ueber einige bei Nervenkranken häufig vorkommende abnorme Mitbewegungen im Fusse und in dem Zehen. Neurolog. Centralblatt, 1 Jan., 1887.

care. During pregnancy an unsuccessful effort was made by the mother to induce an abortion, which she now thinks the cause of the child's condition. Labor came on at term, without any accident. For the first nine months the child is said to have cried most of the time. At no time has there been a history of any illness more than the ordinary complaints of childhood. Since nine months of age, she has been in her present condition of constant muscular rigidity. She is unable to stand without support, but when held up by the arms she can bear most of her weight on the feet, although she says her left leg is "n. g." (no good). Examination showed a dislocation of the left hip, probably congenital. She presents no deformity or muscular atrophy; the muscles are well developed, and even slightly hypertrophied. Her nurse states that at times the muscular rigidity ceases for a minute, and she becomes more limber. The rigidity is greater in the morning and in bad weather. She can neither stand nor sit, but either lies on the floor or is slung by the arms to a spiral spring in the doorway, which she enjoys very much. Any movement, touch, or excitement exaggerates the spasm. At times she has crying and screaming spells. She can move her head very well, and can roll over. She will not stay on her back, and can be put there only with difficulty. If put on her back, she rolls over to the left side with some difficulty, and gets on her belly, which is almost the only voluntary movement, except of the head, which she makes She does this when told to, as well as at other times, but the reverse process, from the belly to the back, cannot be performed. She sleeps on her belly, turning her head to the left, so as to lie on the right side of her face.

During sleep she is very apt to raise the left arm from the bed, letting it fall with such force upon her face as to hurt herself, so that her nurse ties that arm to the side of her crib. At times she gets a little blue about the lips. Except for occasional attacks of diarrhœa she seems well, and makes no complaint. She eats and sleeps well; she has perfect control over her sphincters. Mentally she is said to be unusually bright. Her speech, however, is very indistinct, so that it can be understood only with difficulty. The trouble, however, is purely one of motor inco-ordination, for she talks readily, and always uses the proper words. Her disposition is remarkably sweet, and she is, except when she has a crying spell, of a very sunny and even temper. She has not been taught to read, but her memory is very good. She learns the words of little poems in her picture books on two or three repetitions; she enjoys music, catches the words and airs of the popular songs of the day, and can sing them herself in good tune and time. She knows and can name and distinguish the ordinary colors, takes a lively interest in what is going on about her, remembers me and calls me by name after three months. She enjoys watching the people on the street and the distant landscape, and seems to appreciate the beauty of a sunset. She is docile and obedient. The child is well developed, well nourished, the muscles a trifle hypertrophied. There is dislocation of the left hip. The vision is good, the pupils react to light, and the field of vision and ocular movements are normal. She keeps the mouth open, but can close it, and she does not drool. The tongue can be moved naturally in every direction. She has a habit of raising and lowering the under jaw rather stiffly, and of nodding

the head slowly, especially when talking or excited. She can twist her head and shake it from side to side, but, as a rule, keeps it pretty well back. The body is kept rigid, the arms extended in the crucifixion attitude, her legs rigidly extended, the feet extended, inverted, and in (plantar) flexion,—the spastic position. On lifting her up and requesting her to touch an object, she seems to have a little power to move the arm backward from the plane of the body, but this, and the rolling over on to her belly, are the only voluntary movements, except of the head, which she can make. She never keeps her arms by her side. If anything is put into her hand the fingers close round it tightly, but without any voluntary action on her part—much as a little baby will grasp your finger. The entire body is kept in a ramrod-like rigidity, so that if you want to make her sit up, you must double her up by force, like a spring-bladed jack-knife, and hold her in position, so that she will not straighten out again. This same resistance to passive motion is noticed in all the muscles. At times she is limber enough to be made to bend the hips comfortably to sit in her nurse's lap, but any excitement or touch brings on this ramrod-like rigidity. Any attempt at passive motion is resisted with great strength. At times, however, the limbs seem to move, independently of the will, in a very awkward fashion, although with considerable power. The foot, for instance, is everted, the arms move, the legs are drawn up, but none of these movements can be performed by the will, and they all show marked inco-ordination. The sensation is normal. The muscular spasm was such as to render any attempt to test the reflexes nugatory. The same may be said also of the elec-

trical tests, although it was possible to get a response from the deltoid to the faradic current. The application of the poles elsewhere produced such an exaggeration of the spasm as to prevent the reaction.

In these three cases we have a cerebral infantile paralysis — a double hemiplegia in the last, — with a rather definite group of motor disturbances. The cases differ markedly, of course, from the forms of hemiplegia with late contracture and atrophy which I reported in my former paper.

This form of motor disturbance is unlike athetosis or post-hemiplegic chorea in that the spontaneous movements are rare; the phenomena are excited wholly or chiefly by intended movements, and the characteristic athetoid or choreic movements are absent. It differs moreover from the post-hemiplegic ataxia of Grasset[5] in that other elements beside ataxia go to make up the picture. In Observation I., for instance, we see first a tonic spasm of the paralyzed side, increased by excitement, by efforts to use the affected side or even the other side, thus coming under the heading of a true associated movement. In Observation II. the character of an associated movement is so pronounced that we even find a special form of it described by Strümpell. This tonic spasm is certainly akin to the contracture of ordinary hemiplegia, yet it is, if I may be allowed the expression, more "active" than contracture, and less fixed in its type. In late contracture, there is a passive, fixed shortening of the muscles, which does not vary; this, on the contrary, resembles more the active contraction of voluntary movement. In addition to the tonic

[5] J. Grasset. D'une variété non décrite de phénomène posthémiplégique (Forme hémiataxique). Le Progrés Médical, 13th Nov., 1880.

spasm and the associated movements, there is also, in Observation I. and II., a distinct loss of the power of co-ordination. In Observation III. the spasm predominates; associated movements are thrown so far in the background as almost to escape notice, and the spasm has so far overcome any volitional power over the muscles that inco-ordination is also a subordinate factor.

This symptom-complex — tonic spasm, associated movement, and inco-ordination — although, of course, merely one of Greidenberg's "mixed forms," seems to be a fairly distinct type of post-hemiplegic disturbance of motion. It is not very uncommon, and in my search through the literature of the subject I have found various cases reported under the headings of chorea or athetosis, notably by Fletcher Beach,[6] and also by Clay Shaw'[7]—"imbecility with ataxia." Gowers, moreover, has described a form of slow, mobile spasm following hemiplegia which is closely akin to this. As he first described it,[8] the spasm was involuntary, but it was exaggerated on motion. Since then he has given a fuller description.[9] "The most common form is that in which there is tonic spasm, slowly varying in relative degree in different muscles, and thus causing slow, irregular movements, chiefly conspicuous in the hand, and slow, irregular inco-ordination. From this character it may conveniently be termed 'mobile spasm.' It is commonly conjoined with more or less permanent rigidity, which tends to fix

[6] Fletcher Beach. On Cases of Athetosis. British Med. Journal, 12th June, 1886.

[7] T. Clay Shaw. On Athetosis, or Imbecility with Ataxia. St. Bartholomew's Hospital Reports, ix. 130, 1873.

[8] W.R. Gowers. On "Athetosis" and Post-hemiplegic Disorders of Movement. Medico-Chirurgical Transactions, lix. 219, 1876.

[9] W. R. Gowers. A Manual of Diseases of the Nervous system, ii. 79, 1888.

the limb in a certain posture." This spasm may cease during rest, but it is renewed by any attempt at voluntary movement, which is disordered by the spasm, being rendered ataxic or inco-ordinate. With this slow, mobile spasm are often associated other involuntary movements, which may persist when the limbs are at rest, and may assume the character of athetosis or chorea. This type differs somewhat from the type I have described in being apparently more variable, and in having less associated movement combined with it.

Of the pathology little can be said. Gowers thinks that his "slow, mobile spasm" is especially common in those cases of meningeal hæmorrhage which he calls[10] "cerebral birth palsy," but certainly it cannot be regarded as pathognomonic of a hæmorrhage, as the evidence of difficult parturition is lacking in some cases. I am not disposed to regard this condition as having any localizing character, not even as pointing to a lesion near the thalamus. In Observations I. and II. I am disposed to think the lesion was cortical, but I have seen this tonic spasm with inco-ordination as a transitory symptom at times in cases of ordinary chorea, where the symptoms were unilateral. I think that the most that can be said at present is that this "mixed form" of tonic spasm, inco-ordination, and associated movement, like most if not all of the other forms of post-hemiplegic disturbances of motion, points simply to a lesion of the pyramidal tract which either deranges the initiation of movement in the motor centres, or impairs the conduction of the motor impulse in the nerve fibres.

[10] W. R. Cowers. Clinical Lecture on Birth Palsies. Lancet, 14, 21 April, 1888.

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